Comment

COMMENTS ON A REPORT OF THE AORTO-PULMONARY WINDOW (APW) TYPE I COEXISTING WITH PULMONARY VALVE STENOSIS AND OCCLUDED ARTERIAL DUCT, DETECTED PRENATALLY AT 26 WEEKS OF GESTATION

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SUMMARY

In the article a case of a little APW coincidented with pulmonary stenosis **(the first such description in Poland)** in 26-week-old fetus of a thirty-two-year-old woman, gravida 3, was presented[1].

The reason for a detailed fetal echocardiographic scan was presence of ventricular disproportion in 20 week screening ultrasound. In fetal echo examination in the tertiary care center a 2 mm large APW above the pulmonary valve (type I according to the classification of Mori et al.) and pulmonary valve stenosis (assessed as mild) were detected. **There was no patent ductus arteriosus (DA) visualized during the examination. Its presence was not confirmed neither in this scan nor in subsequent examinations.** Furthermore, an enlarged left ventricle with abundant trabeculation and impairement of function of right ventricle were noted. Digoxin and steroid therapy was administered at 29 weeks of gestation. Subsequent echo at 36 weeks of gestation showed improved function of the RV. Baby girl was delivered by CS at 39 weeks in a good clinical condition. Couple hours later the oxygen saturation level dropped to 80%. The echocardiographic exam in Pediatric Cardiology Department revealed enlarged left atrium appendage. Left ventricle had an increased trabeculation as in the prenatal study. The pulmonary valve was dysplastic (of a narrowed annulus and thickened leaflets) with the pressure gradient of 39,7 mmHg (3,15 m/s). The neonate was discharged in good clinical condition presenting satisfactory weight gain.

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Comment of Prof. A. Rudziński:

Aortopulmonary window is a rare congenital heart defect. Its prevalence is about 0,1-0,3 % of all CHD. The APW is a result of developmental deficiency of the septum between the aorta and pulmonary artery. The spiral septum dividing the great arteries is built by fusing the truncal septum arising cranially from the semilunar leaflets level and the aorto-pulmonary septum, descending caudally oppositely to the pulmonary branches confluence. In 48 % of all cases APW accurs as isolated CHD, in 52 % it is accompanied by other cardiac anomaly (PDA, IAA, CoAo, TGA, TOF, VSD, ASD)[2]. Predominantly (75%) it is coincidented with the PDA, and frequently with the IAA type A[3]. Before the surgical repair the coexisting anomalous origin of coronary arteries (arising from pulmonary artery) should be ruled out.

The first case of the APW was described by Elliotson in 1830[4], whereas the first prenatal echocardiographic diagnosis was made by Collinet et al. in 2002[5]. One of the first classifications depending on size and location

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of APW was estalished by Mori et al. It distinguishes three types: I – proximal, II – distal and III – complete[6]. The APW described in this article would be classified as type I. Small restrictive window occurs rather rarely (in about 10 % of all cases) and it is usually detected in the late infancy because of the oligosymptomaticity (mainly a murmur upon auscultation). Large APW give symptoms earlier, manifesting its presence by congestive heart failure besides the murmur. Symptoms appear earlier in ductal-dependent defects (IAA type A or supraductal CoAo) as a result of a postnatal restriction of the DA. In children presenting a high pulmonary resistance symptoms can be unnoticed until pulmonary hypertension becomes persistent. At this time symptoms characteristic for Eisenmenger syndrome appear (cyanosis, fatigue, syncopes on exertion, the murmur of the tricuspid and pumonary valves regurge, increased accentuation of the II heart sound).

In fetus with isolated APW the echocardiographic exam should reveal right-to-left shunt and potential dilatation of the aorta. These features despite the small size of the APW were present in this case. It could be a result of an absent DA and supra-systemic pressure in pulmonary circulation leading to this shunt (observed in spite of pulmonary valve stenosis). This pressure overload could be the reason for the right ventricle disfunction. Presence of APW in this case interacted preferably decompressing the pulmonary circulation, in which increased pressure was suspected in the absence of DA. The flow through pulmonary circulation in fetal life is meager (about 10 % of cardiac output – CO – as a result of high resistance). Major part (90%) of CO is directed through the DA to the systemic circulation.

In my opinion, the abundant trabeculation in the left ventricle was a result of the left ventricular non-compaction, leading in extreme conditions to cardiomyopathy, congestive heart failure, arrythmias and thromboembolic incidents. The pulmo-arterial shunt through a small APW could not be a factor of volume overload of the left ventricle (even large APWs are not causing any alternations in fetuses). I found the suggestions of influence of steroid therapy on restriction of the DA unjustified, because the DA was not observed in the previously performed examinations. There is also no evidence of an effect of maternal steroid-therapy on DA closing in literature. It is known that maternal administration of steroids causes contriction of fetal DA and physiologically increases the cortisol level in a late pregnancy, which plays a role in its postnatal closing[7].

The accurate assessement of pulmonary stenosis in presence of even small APW in Doppler echocardiographic exam is illusory as a result of a tendency to equalize pressure between the great arteries. I find it hard to comment on appendages assessment exclusively on a basis of the exam description. Suggestions that the IVC was connected to the LA also seem doubtful. In such cases severe oxygen desaturations in systemic circulation occur, which was not observed in this case.

In conclusion, I would like to congratulate the prenatal observation of the APW in an unusual coincidence (small APW+PVS) additionally with absent PDA resulting in supra-systemic pressure in pulmonary circulation, which is ameliorated by present APW. Its absence could lead to severe and irreversible alterations in pulmonary circulation.

References:

1. *Więckowska K, Węgrzynowski J, Zych - Krekora K, Kwiatkowska J, Słodki M, Respondek-Liberska M. Antenatal echocardiographic diagnosis of an aortopulmonary window combined with pulmonary stenosis in neonate. A case report. Prenat Cardio. 2015;5:33-37*

2. *Yen Ho S., Gerlis L M, Andreson C et al. The morphology of aortopulmonary windows with regard to their classification and morphogenesis. Cardiol Young 1994;4:146-55*

3. *Mert M, Paker T, Akcevin A, et al. Diagnosis, management, and results of treatment for aortopulmonary window. Cardiol Young. 2004;14:506-11.*

4. *Elliotson J. Case of malformation of the pulmonary artery from the aorta. Lancet 1830;31:247-8*

5. *Collinet P et al. Prenatal diagnosis of an aorto-pulmonary window by fetal echocardiography Fetal Diagn Ther 2002; 17:302-307.*

6. *Mori K, Ando M, Ishikawa S, Imai Y. Distal type of aorto-pulmonary window. Report of 4 cases. Brit Heart J 1973;40:681-9.*

7. *Clyman RI. Mechanism regulating the ductus arteriosus. Biol Neonate 2006;89:330-5*